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Spontaneous intracerebral hemorrhage in hemophiliacs—A treatment dilemma

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ABSTRACT

BACKGROUND: Spontaneous Intracerebral hemorrhage is a rare and challenging condition to treat, especially in haemophiliacs. With their innate predilection to bleed following trivial trauma, surgical management of such cases have been individualised and no treatment protocols exist. Often they are managed with craniotomy and evacuation under cover of AHF.

DISCUSSION: Here we discuss the nuances, clinical and treatment dilemma that one faces while treating a patient, diagnosed with haemophilia, with spontaneous subdural haematoma. While routine management strategy would be a craniotomy and evacuation of the haematoma, in our case, we chose to closely monitor the patient under cover of AHF, to convert the acute haematoma into a chronic subdural hematoma. We then managed it with a burr hole evacuation under cover of antihemophilic factor.

CONCLUSION: Treatment protocols are hard to formulate in such rare entities, however we would like to recommend the following; conservative management of an acute subdural to convert it into a chronic subdural when there are no financial constraints, there by obviating the complication of rebleed following craniotomy.

1. Introduction

Spontaneous Intracerebral hemorrhage is a rare and challenging condition to treat, especially in haemophiliacs. The risk-benefit ratio that tilts the balance between conservative and surgical management is very subtle. Surgical treatment bears the risk of rebleed, whereas conservative management runs the risk of rapid deterioration and high treatment cost [1]. Here we discuss the nuances, clinical and treatment dilemma that one faces while treating a patient, diagnosed with haemophilia, with spontaneous right subdural haematoma. Here we present our clinical vignette.

2. Case report

A 32 year old doctor, diagnosed case of Christmas disease, presented to the triage with sudden onset, progressive drowsiness and altered sensorium since the previous night. On further assessment, he revealed that he had holocranial headache since the last 1 week which was associated with vomiting. He had no prior history of trauma. On examination he had bradycardia, pulse rate of 60/min and neurological examination revealed a disoriented, drowsy patient, who was obeying simple commands with no paucity of movements. Pupils were equal and reacting well. A preliminary computed tomographic scan of the brain (CT Brain) revealed an acute right fronto-temporo-parietal acute subdural haematoma with mass effect and midline shift of 1.6cms. The basal cisterns were partially effaced and there was significant cerebral oedema.

Haematological workup was requested and the reports showed an elevated Activated plasma thromboplastin time (aPTT—55 s) with low Factor VIII levels (4.9%). Prothrombin time and INR were within normal limits.

The dilemma in the treatment was to weigh the benefits versus the risks of performing a craniotomy for evacuation of the acute SDH as compared to conservative management with correction of coagulation profile followed by burr hole and evacuation.

Since the patient was neurologically stable a clinical judgement was made, after counselling with his relatives, to attempt at conservative management and correction of coagulation factors under continuous neuromonitoring in our Neuro Intensive Care Unit.

After consulting with the inhouse haematopathologist it was decided to start the patient on Anti Hemophilic Factor (AHF) to raise the desired level to 100 IU/dl. He was infused with the weight calculated dose for the following three days and his Factor VIII levels reached 153%. Follow up CT Scan brain showed no significant change in the haematoma size however the patient appeared to be significantly drowsy and his bradycardia had worsened (45 beats/min) on day 5 of admission. The patient was obeying simple commands and the relatives were counselled regarding the same

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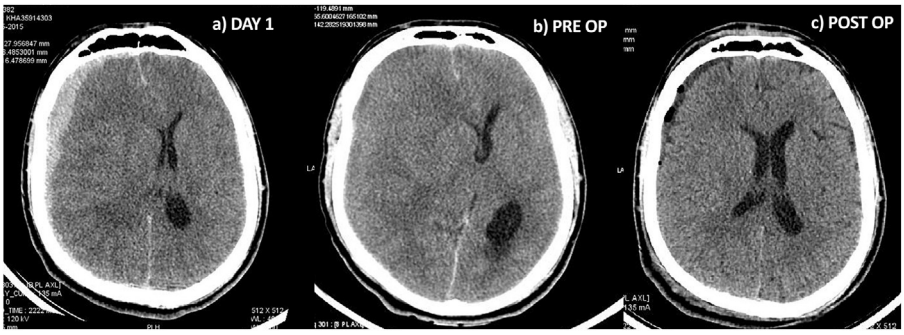

Fig. 1. a) CT Scan on admission b) Pre Operative CT Scan c) Post OP CT Scan.

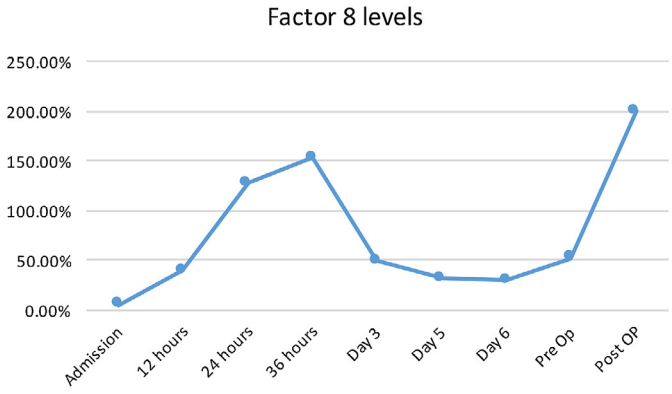

Fig. 2. Factor 8 levels during the course of treatment.

and decided to continue conservative line of management with AHF (Figs. 1 and 2).

Decongestants and AHF were continued and on the 7th day post admission he developed pupillary asymmetry, with deterioration of sensorium. CT Scan done revealed a right fronto-temporo-parietal subacute subdural hematoma with midline shift. Patient attenders were re-counselled and explained the need for immediate surgical evacuation. He was taken up for emergency burr hole and evacuation of haematoma under cover of AHF under general anesthesia. Preoperative and postoperative transfusions of AHF was done to maintain the AHF levels. Postoperatively, within 6 h patient was alert, active and oriented, CT brain showed complete evacuation of hematoma and resolution of midline shift. Factor 8 infusion was continued at maintenance doses for the next 7 days and he was discharged with no complications and is on regular follow-up.

3. Discussion

Spontaneous intracranial hemorrhage is a rare complication of hemophilia, with frequency of about 2.2–7.8% [2], and a mortality of 34% [3]. It is a life threatening complication and needs emergent neurosurgical intervention. Small volume bleeds with insignificant mass effect and midline shift may be managed conservatively with AHF and close neurological monitoring, allowing spontaneous resolution of hematoma [4]. Larger bleeds, like the current case, should be operated under the cover of AHF. In our case we were able to successfully convert the acute subdural hematoma to a chronic subdural hematoma under cover of decongestants and AHF. Only once the patient was neurologically obtunded, surgical evacuation was contemplated. This avoided the need for craniotomy and the relatively high risk of rebleed [5].

Management of our cases with AHF was based on guidelines issued by the World Federation of Hemophilia [6]. Factor VIII concentrate, Factor IX concentrate, Cryoprecipitate, Fresh Frozen Plasma (FFP), Desmopressin, Tranexemic Acid, Epsilon AminoCaproic acid are the products available for the management of Hemophilia and bleeding manifestations. In our case we used Factor VIII concentrates with reserve of FFP and Cryoprecipitate. Each unit of Factor VIII infused per kilogram of body weight increases the plasma level of factor VIII by 2 IU/dl [7]. Factor VIII has a half life of approximately 8–12 h. Factor VIII requirement was calculated using the formula Weight in Kilogram × desired concentration (IU/dl) × 0.5. The prescribed level of factor VIII levels for neurosurgical emergencies is 100 IU/dl and the calculated dose was 3800 IU (76 kg × 100 IU/dl × 0.5). This dose of 3800 IU was infused intravenously twice daily for 3 days. The dose of factor VIII was then reduced to achieve a maintenance concentration of 50IU (76 kg × 50 IU/dl × 0.5) i.e 1900 IU was infused over the next 5 days till he deteriorated and was taken up for surgery. Prior to the surgical procedure a bolus dose of 3800 IU was given intravenously and a pre op factor VIII level of 200%. The same dose was continued for 3 days postoperatively and then tapered to a level of 50 IU/dl for the next 5 days and 25 IU/dl for 3 days thereafter.

Surgical indications for the management of large intracranial hematomas in Hemophiliacs have been no different from non hemophiliacs in the past [7]. We however successfully converted an acute subdural hematoma to chronic subdural hematoma and managed it by burr hole evacuation. The decision and favourable outcome was possible in view of the good GCS the patient presented with. This technique not only significantly reduced the morbidity of craniotomy and rebleed [5] but also gave a good functional outcome (Glasgow Outcome score of 5). The drawbacks of the above management technique included the high costs of AHF and the ambiguity of constant deterioration of the patient.

4. Conclusion

Our clinical experience has enlightened us regarding the solution to a few dilemmas one faces while treating an acute subdural in a haemophiliac. We would like to recommend the following; conservative management of an acute subdural to convert it into a chronic subdural when there are no financial constraints. This reduces the obvious yet unavoidable complication of rebleed and the morbidity following craniotomy.

Conflicts of interest

No.

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Ethical approval

NA.

Author contribution

Dr Ajay Hegde – writing the paper.
Dr Rajesh Nair – data collection and design.
Dr Sunil Updhyaya – Operating surgeon.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Guarantor

Dr Rajesh P. Nair.

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